Fig.1

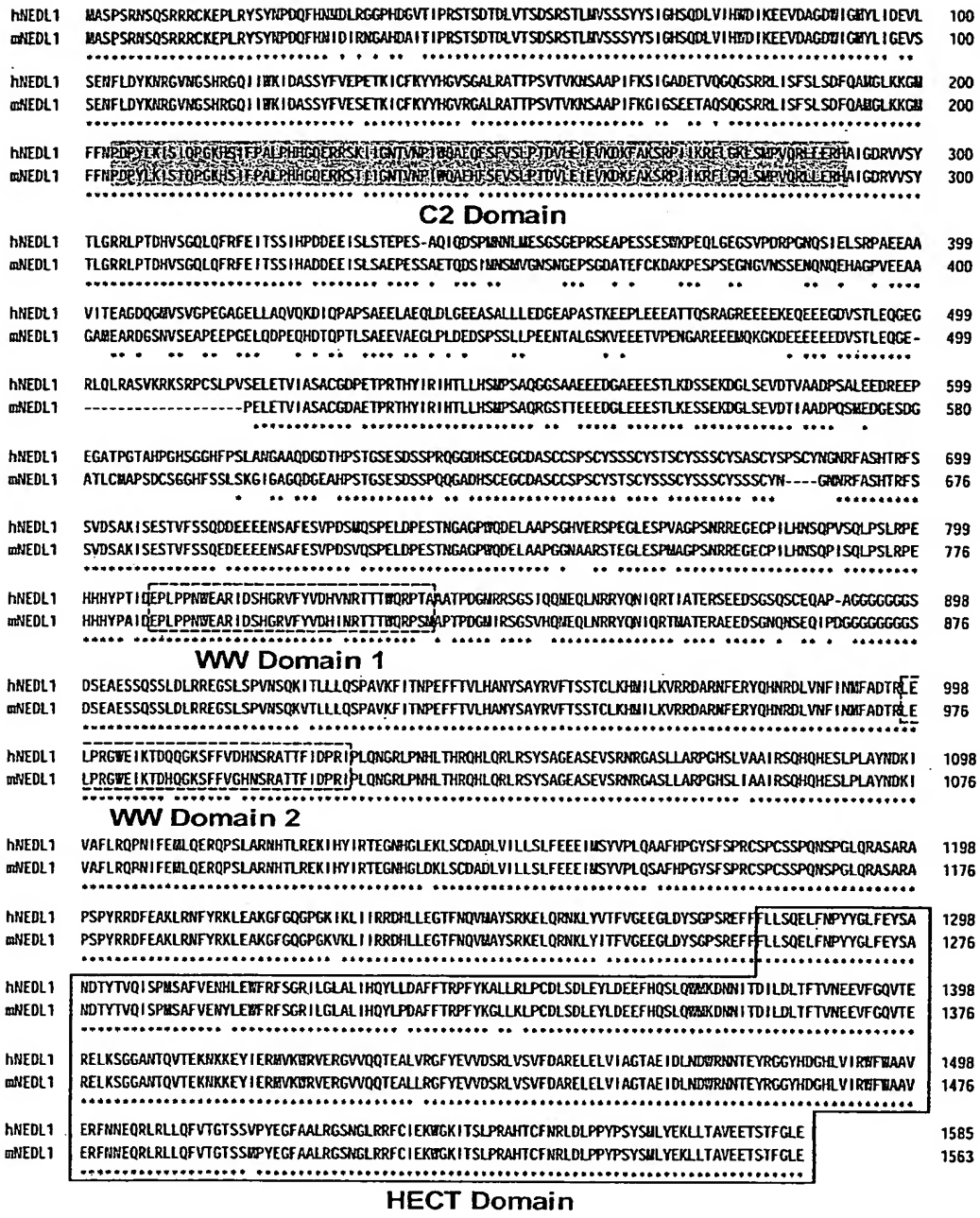


Fig.2

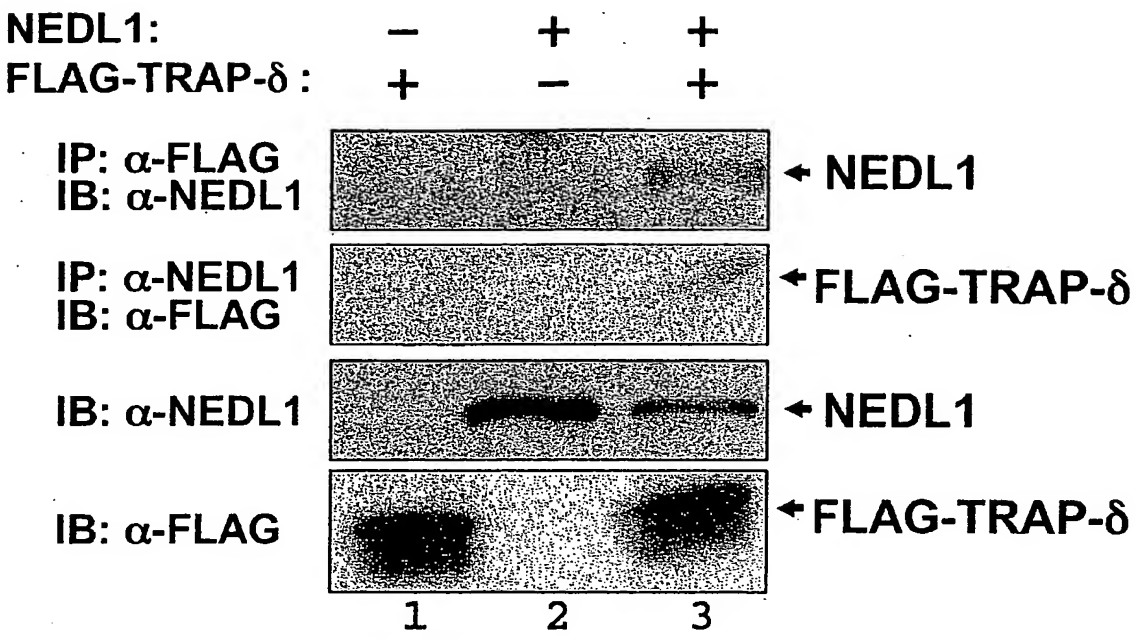

NEDL1: – + +
FLAG-TRAP-δ : + – +
IP: α-FLAG
IB: α-NEDL1
NEDL1
IP: α-NEDL1
IB: α-FLAG
FLAG-TRAP-δ
IB: α-NEDL1
NEDL1
IB: α-FLAG
FLAG-TRAP-δ
1 2 3

Fig.3

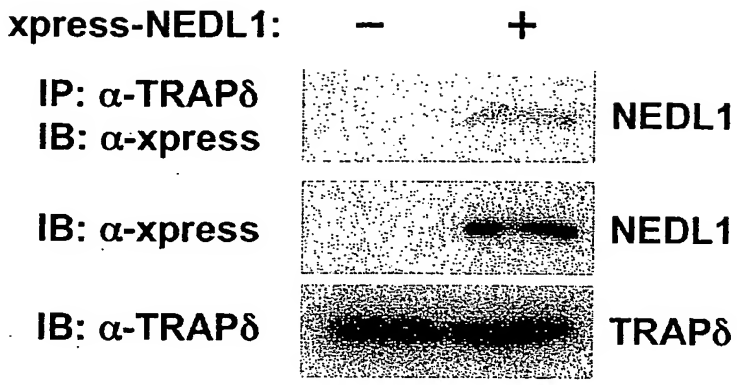

xpress-NEDL1: − +
IP: α-TRAPδ
IB: α-xpress
NEDL1
IB: α-xpress
NEDL1
IB: α-TRAPδ
TRAPδ

Fig.4

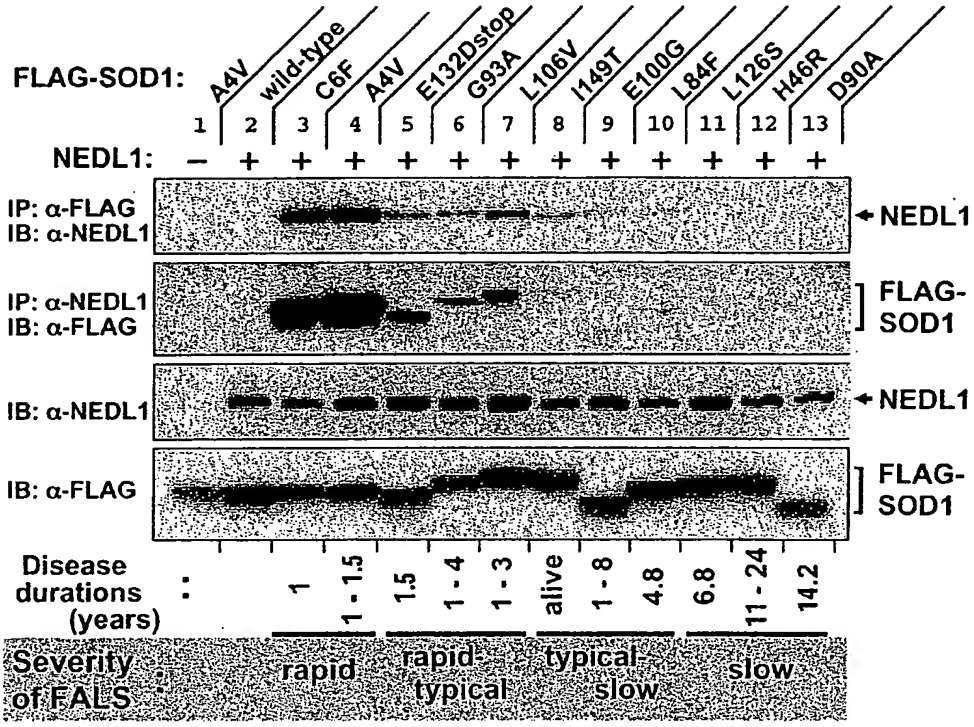

FLAG-SOD1:
A4V
wild-type
C6F
A4V
E132Dstop
G93A
L106V
I149T
E100G
L84F
L126S
H46R
D90A
1 2 3 4 5 6 7 8 9 10 11 12 13
NEDL1: – + + + + + + + + + + + +
IP: α-FLAG
IB: α-NEDL1
NEDL1
IP: α-NEDL1
IB: α-FLAG
FLAG-SOD1
IB: α-NEDL1
NEDL1
IB: α-FLAG
FLAG-SOD1
Disease durations (years)
: 1 1 - 1.5 1.5 1 - 4 1 - 3 alive 1 - 8 4.8 6.8 11 - 24 14.2
Severity of FALS
rapid
rapid-typical
typical
slow
slow

Fig.5

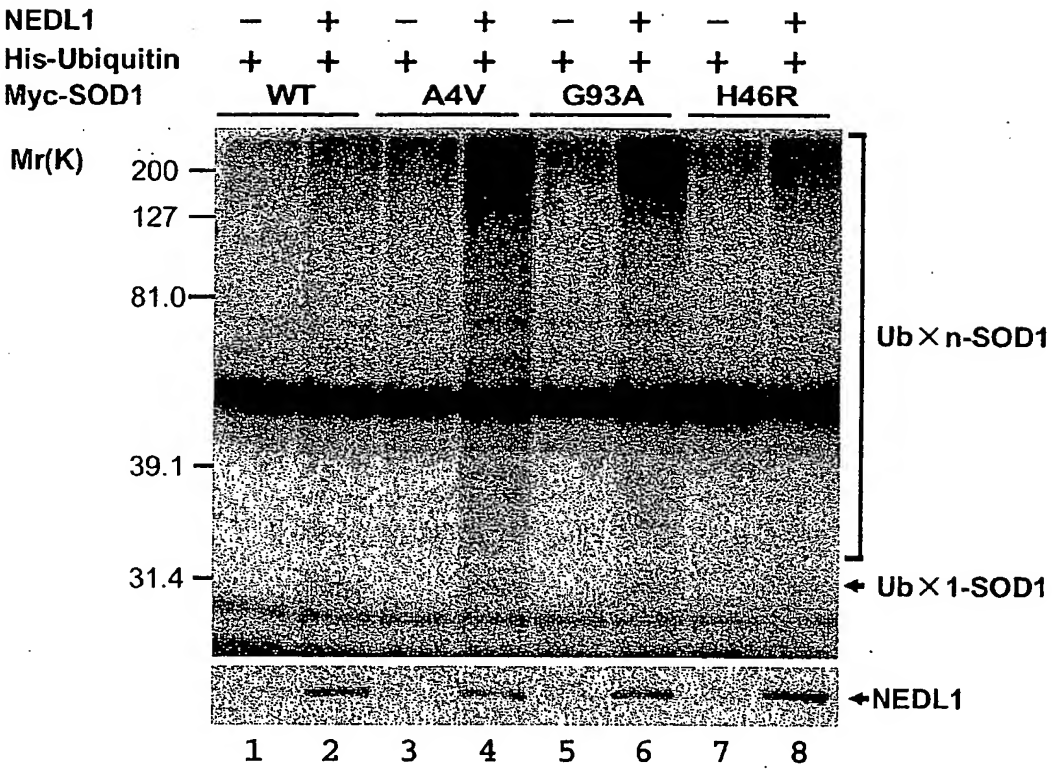

NEDL1
His-Ubiquitin
Myc-SOD1
WT
A4V
G93A
H46R
Mr(K)
200
127
81.0
39.1
31.4
Ub×n-SOD1
Ub×1-SOD1
NEDL1
1 2 3 4 5 6 7 8

Fig.6

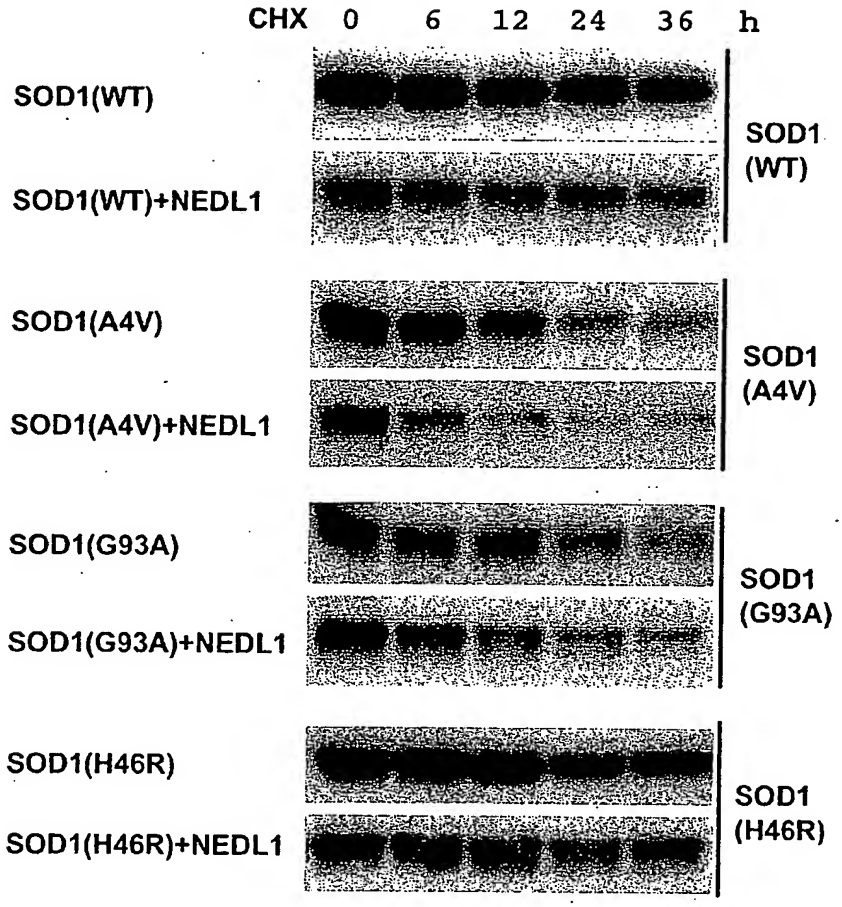

CHX 0 6 12 24 36 h
SOD1(WT)
SOD1(WT)+NEDL1
SOD1 (WT)
SOD1(A4V)
SOD1(A4V)+NEDL1
SOD1 (A4V)
SOD1(G93A)
SOD1(G93A)+NEDL1
SOD1 (G93A)
SOD1(H46R)
SOD1(H46R)+NEDL1
SOD1 (H46R)

Fig.7

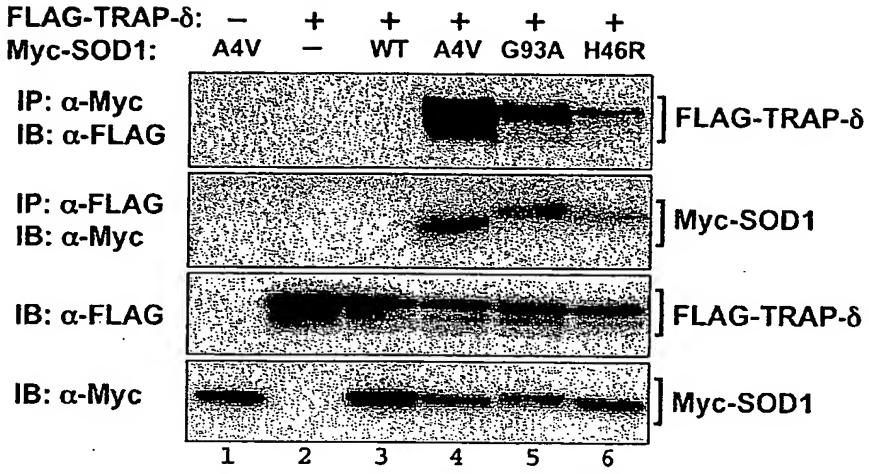

FLAG-TRAP-δ: – + + + + +
Myc-SOD1: A4V – WT A4V G93A H46R
IP: α-Myc
IB: α-FLAG
FLAG-TRAP-δ
IP: α-FLAG
IB: α-Myc
Myc-SOD1
IB: α-FLAG
FLAG-TRAP-δ
IB: α-Myc
Myc-SOD1
1 2 3 4 5 6

Fig.8

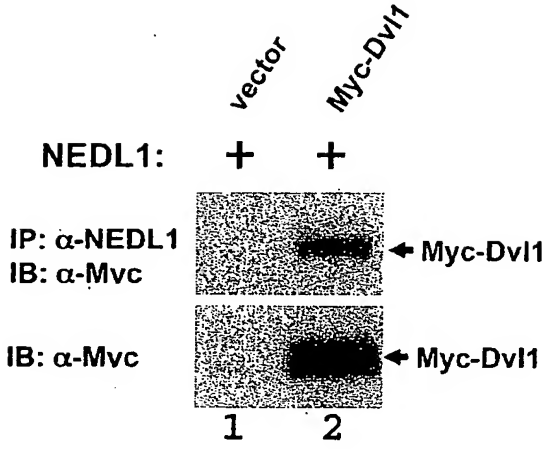

vector
Myc-Dvl1
NEDL1:
+
+
IP: α-NEDL1
IB: α-Mvc
Myc-Dvl1
IB: α-Mvc
Myc-Dvl1
1
2

Fig.9

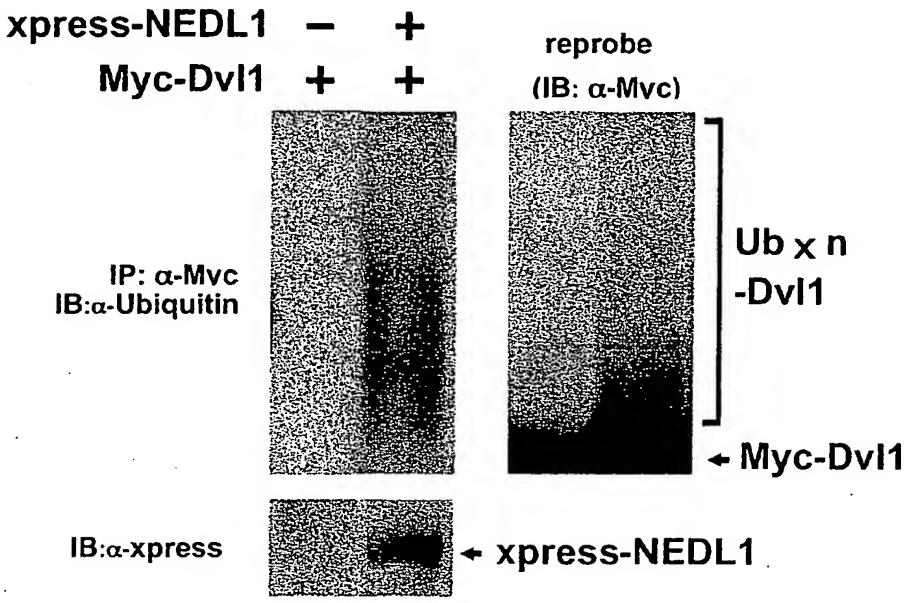

xpress-NEDL1 – +
Myc-Dvl1 + +
reprobe
(IB: α-Mvc)
IP: α-Mvc
IB:α-Ubiquitin
Ub x n
-Dvl1
Myc-Dvl1
IB:α-xpress
xpress-NEDL1

Fig.10

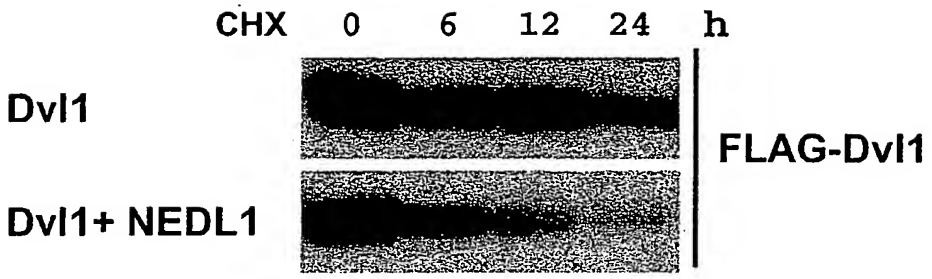
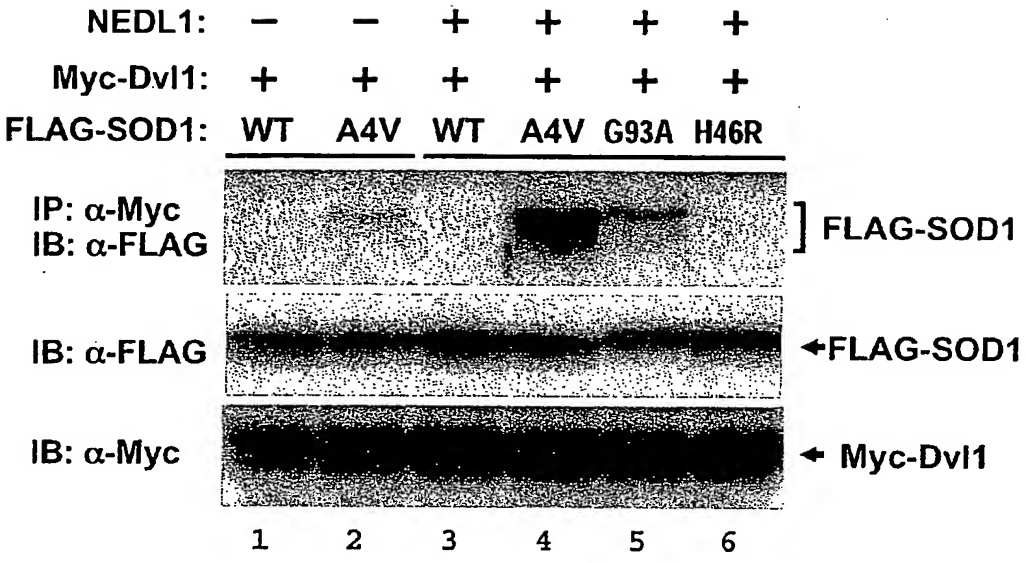


Fig.11


NEDL1: − − + + + +
Myc-Dvl1: + + + + + +
FLAG-SOD1: WT A4V WT A4V G93A H46R
IP: α-Myc
IB: α-FLAG
FLAG-SOD1
IB: α-FLAG
FLAG-SOD1
IB: α-Myc
Myc-Dvl1
1 2 3 4 5 6

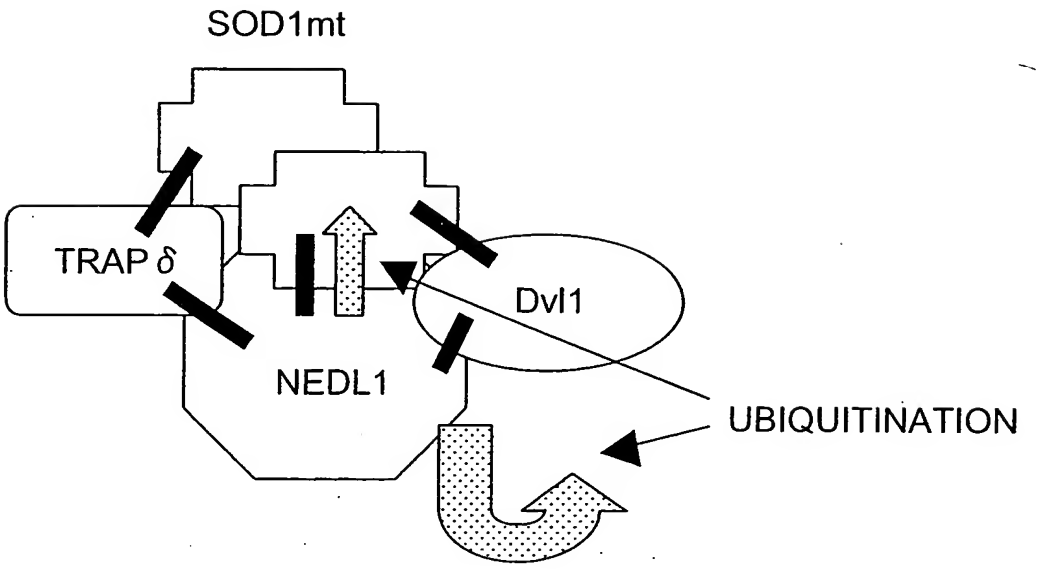



Fig.12